Message from the Editor

1 Robert A. Fishman, MD

Editorials

- 3 Mitochondrial Myopathy in the Elderly: Exaggerated Aging in the Pathogenesis of Disease Jerry R. Mendell, MD
- 5 Clinical Outcomes and Documentation of Partial Beneficial Effects of Immunotherapy for Multiple Sclerosis John N. Whitaker, MD, Galen W. Mitchell, MD, and Gary R. Cutter, PhD

Neurological Progress

7 The Interferons: Biological Effects, Mechanisms of Action, and Use in Multiple Sclerosis Bianca Weinstock-Guttman, MD, Richard M. Ransohoff, MD, R. Philip Kinkel, MD, and Richard A. Rudick, MD

Original Articles

- 16 Late-Onset Mitochondrial Myopathy Wendy Johnston, MD, George Karpati, MD, Stirling Carpenter, MD, Douglas Arnold, MD, and Eric A. Shoubridge, PhD
- 24 Ragged Red Fibers in Normal Aging and Inflammatory Myopathy Ziad Rifai, MD, Stephen Welle, PhD, Cornelia Kamp, BS, and Charles A. Thornton, MD
- 30 Low-Dose (7.5 mg) Oral Methotrexate Reduces the Rate of Progression in Chronic Progressive Multiple Sclerosis

 Donald E. Goodkin, MD, Richard A. Rudick, MD, Sharon VanderBrug Medendorp, MPH,

 Michele M. Daughtry, BA, Kathleen M. Schwetz,

 Jill Fischer, PhD, and Carolyn Van Dyke, MD
- 41 Myopathy and Paraproteinemia with Serum IgM Binding to a High-Molecular-Weight Muscle Fiber Surface Protein M. T. Al-Lozi, MD, A. Pestronk, MD, W. C. Yee, MD, and N. Flaris, MD
- 47 Histometric Effects of Ciliary Neurotrophic Factor in Wobbler Mouse Motor Neuron Disease Ken Ikeda, MD, Vivien Wong, PhD, Thomas H. Holmlund, MD, Tom Greene, PhD, Jesse M. Cedarbaum, MD, Ronald M. Lindsay, PhD, and Hiroshi Mitsumoto, MD
- 55 Circulating, Soluble Adhesion Proteins in Cerebrospinal Fluid and Serum of Patients with Multiple Sclerosis: Correlation with Clinical Activity Paula Dore-Duffy, PhD, Walter Newman, PhD, Roumen Balabanov, MD, Robert P. Lisak, MD, Elizabeth Mainolfi, MS, Robert Rothlein, PhD, and Margaret Peterson, PhD

- 63 Elevated Mast Cell Tryptase in Cerebrospinal Fluid of Multiple Sclerosis Patients
 J. J. Rozniecki, S. L. Hauser, M. Stein, R. Lincoln, and T. C. Theobarides
- 67 The Trends in Incidence of Primary Brain Tumors in the Population of Rochester, Minnesota

 Kurupath Radhakrishnan, MD, Bahram Mokri, MD, Joseph E. Parisi, MD, W. Michael O'Fallon, PhD, Jayasimha Sunku, MD, and Leonard T. Kurland, MD, DrPH
- 74 Regional Cerebral Blood Flow Changes in Motor Cortical Areas after Transient Anesthesia of the Forearm Norihiro Sadato, MD, Thomas A. Zeffiro, MD, PhD, Gregory Campbell, PhD, Junji Konishi, MD, Hiroshi Shibasaki, MD, and Mark Hallett, MD
- 82 Tumor Necrosis Factor-α Messenger RNA
 Expression in Patients with Relapsing-Remitting
 Multiple Sclerosis Is Associated with Disease
 Activity
 P. Rieckmann, MD, M. Albrecht, BA, B. Kitze, MD,
 T. Weber, MD, H. Tumani, MD, A. Broocks, MD,
 W. Lüer, MD, A. Helwig, MD, and S. Poser, MD
- 89 Experimental Ischemic Neuropathy: Salvage with Hyperbaric Oxygenation
 Mikibiro Kihara, MD, Philip G. McManis, MD, James D. Schmelzer, BS, Yoriko Kihara, MD, and Phillip A. Low, MD
- 95 Comparative Study of Selegiline Plus L-Dopa-Carbidopa Versus L-Dopa-Carbidopa Alone in the Treatment of Parkinson's Disease Timothy Brannan, MD, and Melvin D. Yahr, MD
- 99 Genetic Identity of Fukuyama-type Congenital Muscular Dystrophy and Walker-Warburg Syndrome Tatsushi Toda, MD, Mieko Yoshioka, MD, Yutaka Nakahori, MD, Ichiro Kanazawa, MD, Yusuke Nakamura, MD, and Yasuo Nakagome, MD
- Amyotrophic Lateral Sclerosis Immunoglobulins Increase Ca²⁺ Currents in a Motoneuron Cell Line Dennis R. Mosier, MD, PhD, Pietro Baldelli, PhD, Osvaldo Delbono, MD, PhD, R. Glenn Smith, MD, PhD, Maria E. Alexianu, MD, Stanley H. Appel, MD, and Enrico Stefani, MD, PhD

Brief Communications

10 The CYP2D6B Mutant Allele Is Overrepresented in the Lewy Body Variant of Alzheimer's Disease T. Saitob, PhD, Y. Xia, MS, X. Chen, MD, E. Masliah, MD, D. Galasko, MD, C. Shults, MD, L. J. Thal, MD, L. A. Hansen, MD, and R. Katzman, MD

Message from the Editor

1 Robert A. Fishman, MD

Editorials

- 3 Mitochondrial Myopathy in the Elderly: Exaggerated Aging in the Pathogenesis of Disease Jerry R. Mendell, MD
- 5 Clinical Outcomes and Documentation of Partial Beneficial Effects of Immunotherapy for Multiple Sclerosis John N. Whitaker, MD, Galen W. Mitchell, MD, and Gary R. Cutter, PhD

Neurological Progress

7 The Interferons: Biological Effects, Mechanisms of Action, and Use in Multiple Sclerosis Bianca Weinstock-Guttman, MD, Richard M. Ransohoff, MD, R. Philip Kinkel, MD, and Richard A. Rudick, MD

Original Articles

- 16 Late-Onset Mitochondrial Myopathy Wendy Johnston, MD, George Karpati, MD, Stirling Carpenter, MD, Douglas Arnold, MD, and Eric A. Shoubridge, PhD
- 24 Ragged Red Fibers in Normal Aging and Inflammatory Myopathy Ziad Rifai, MD, Stephen Welle, PhD, Cornelia Kamp, BS, and Charles A. Thornton, MD
- 30 Low-Dose (7.5 mg) Oral Methotrexate Reduces the Rate of Progression in Chronic Progressive Multiple Sclerosis

 Donald E. Goodkin, MD, Richard A. Rudick, MD, Sharon VanderBrug Medendorp, MPH,

 Michele M. Daughtry, BA, Kathleen M. Schwetz,

 Jill Fischer, PhD, and Carolyn Van Dyke, MD
- 41 Myopathy and Paraproteinemia with Serum IgM Binding to a High-Molecular-Weight Muscle Fiber Surface Protein M. T. Al-Lozi, MD, A. Pestronk, MD, W. C. Yee, MD, and N. Flaris, MD
- 47 Histometric Effects of Ciliary Neurotrophic Factor in Wobbler Mouse Motor Neuron Disease Ken Ikeda, MD, Vivien Wong, PhD, Thomas H. Holmlund, MD, Tom Greene, PhD, Jesse M. Cedarbaum, MD, Ronald M. Lindsay, PhD, and Hiroshi Mitsumoto, MD
- 55 Circulating, Soluble Adhesion Proteins in Cerebrospinal Fluid and Serum of Patients with Multiple Sclerosis: Correlation with Clinical Activity Paula Dore-Duffy, PhD, Walter Newman, PhD, Roumen Balabanov, MD, Robert P. Lisak, MD, Elizabeth Mainolfi, MS, Robert Rothlein, PhD, and Margaret Peterson, PhD

- 63 Elevated Mast Cell Tryptase in Cerebrospinal Fluid of Multiple Sclerosis Patients
 J. J. Rozniecki, S. L. Hauser, M. Stein, R. Lincoln, and T. C. Theobarides
- 67 The Trends in Incidence of Primary Brain Tumors in the Population of Rochester, Minnesota

 Kurupath Radhakrishnan, MD, Bahram Mokri, MD, Joseph E. Parisi, MD, W. Michael O'Fallon, PhD, Jayasimha Sunku, MD, and Leonard T. Kurland, MD, DrPH
- 74 Regional Cerebral Blood Flow Changes in Motor Cortical Areas after Transient Anesthesia of the Forearm Norihiro Sadato, MD, Thomas A. Zeffiro, MD, PhD, Gregory Campbell, PhD, Junji Konishi, MD, Hiroshi Shibasaki, MD, and Mark Hallett, MD
- 82 Tumor Necrosis Factor-α Messenger RNA
 Expression in Patients with Relapsing-Remitting
 Multiple Sclerosis Is Associated with Disease
 Activity
 P. Rieckmann, MD, M. Albrecht, BA, B. Kitze, MD,
 T. Weber, MD, H. Tumani, MD, A. Broocks, MD,
 W. Lüer, MD, A. Helwig, MD, and S. Poser, MD
- 89 Experimental Ischemic Neuropathy: Salvage with Hyperbaric Oxygenation
 Mikibiro Kihara, MD, Philip G. McManis, MD, James D. Schmelzer, BS, Yoriko Kihara, MD, and Phillip A. Low, MD
- 95 Comparative Study of Selegiline Plus L-Dopa-Carbidopa Versus L-Dopa-Carbidopa Alone in the Treatment of Parkinson's Disease Timothy Brannan, MD, and Melvin D. Yahr, MD
- 99 Genetic Identity of Fukuyama-type Congenital Muscular Dystrophy and Walker-Warburg Syndrome Tatsushi Toda, MD, Mieko Yoshioka, MD, Yutaka Nakahori, MD, Ichiro Kanazawa, MD, Yusuke Nakamura, MD, and Yasuo Nakagome, MD
- Amyotrophic Lateral Sclerosis Immunoglobulins Increase Ca²⁺ Currents in a Motoneuron Cell Line Dennis R. Mosier, MD, PhD, Pietro Baldelli, PhD, Osvaldo Delbono, MD, PhD, R. Glenn Smith, MD, PhD, Maria E. Alexianu, MD, Stanley H. Appel, MD, and Enrico Stefani, MD, PhD

Brief Communications

10 The CYP2D6B Mutant Allele Is Overrepresented in the Lewy Body Variant of Alzheimer's Disease T. Saitob, PhD, Y. Xia, MS, X. Chen, MD, E. Masliah, MD, D. Galasko, MD, C. Shults, MD, L. J. Thal, MD, L. A. Hansen, MD, and R. Katzman, MD

- 112 Chronic Relapsing Axonal Neuropathy: A First Case Report
 Elisabeth Chroni, MD, Susan M. Hall, DSc, and Richard A. C. Hughes, FRCP
- 115 Neuroprotective Effects of Gangliosides May Involve Inhibition of Nitric Oxide Synthase Ted M. Dawson, MD, PhD, Kenneth Hung, BS, Valina L. Dawson, PhD, Joseph P. Steiner, PhD, and Solomon H. Snyder, MD
- Apolipoprotein E Genotypes and Alzheimer's Disease in a Community Study of Elderly African Americans
 H. C. Hendrie, MBChB, K. S. Hall, PhD,
 S. Hui, PhD, F. W. Unverzagt, PhD, C. E. Yu, PhD,
 D. K. Lahiri, PhD, A. Sahota, PhD, M. Farlow, MD,
 B. Musick, C. A. Class, MD, A. Brashear, MD,
 V. E. Burdine, MD, B. O. Osuntokun, MD,
 A. O. Ogunniyi, MD, O. Gureje, MD,
 O. Baiyewu, MD, and G. D. Schellenberg, PhD
- 120 Drenching Sweats as an Off Phenomenon in Parkinson's Disease: Treatment and Relation to Plasma Levodopa Profile Jacob I. Sage, MD, and Margery H. Mark, MD
- 123 Temporal Lobe Epilepsy Caused by Domoic Acid Intoxication: Evidence for Glutamate Receptor-mediated Excitotoxicity in Humans Fernando Cendes, MD, Frederick Andermann, MD, FRCP(C), Stirling Carpenter, MD, Robert J. Zatorre, PhD, and Neil R. Cashman, MD

History of Neurology

127 Staggering Gait in Medical History Francis Schiller, MD

Letters

- 136 PMP22 Expansion in CMT1a Neuropathy C. O. Hanemann, MD, G. Stoll, MD, and H. W. Müller, PbD
- 136 The Influence of Apolipoprotein E Isotypes on Alzheimer's Disease Pathology in 40 Cases of Down's Syndrome

 Thomas Wisniewski, MD, Laura Morelli, PhD,
 J. Wegiel, VMD, PhD, Efrat Levy, PhD,
 H. M. Wisniewski, MD, PhD,
 and Blas Frangione, MD, PhD
- 138 Antagonist Activity in Cerebellar Hypermetria Barbara Wild, MD
- 138 Reply
 M. Manto, MD, E. Godaux, MD, and J. Jacquy, MD
- 139 A Clue to the Pathogenesis of Dopa-responsive Dystonia Yoshiaki Furukawa, MD, Yoshikuni Mizuno, MD, Katsunori Nishi, MD, and Hirotaro Narabayashi, MD

Regular Departments

140 Corrections

NO 2 FEBRUARY 1995

Editorials

- 141 Outcome Research in Neurology: Incorporating Health-related Quality of Life Orrin Devinsky, MD
- 143 What is the Pathogenesis of Human T-Cell Lymphotropic Virus Type I-associated Myelopathy/Tropical Spastic Paraparesis? Per Höllsberg, MD, and David A. Hafler, MD

Neurological Progress

146 Basic Mechanisms of Generalized Absence Seizures O. Carter Snead III, MD

Original Articles

158 Outcome Assessment for Epilepsy Surgery:
The Impact of Measuring Health-related Quality
of Life
Barbara G. Vickrey, MD, MPH, Ron D. Hays, PhD,
Jerome Engel, Jr, MD, PhD, Karen Spritzer, BS,
William H. Rogers, PhD, Rebecca Rausch, PhD,

Jason Graber, BS, and Robert H. Brook, MD, ScD

- 167 Detection of Human T-Lymphotropic Virus
 Type I (HTLV-I) Tax RNA in the Central
 Nervous System of HTLV-I-associated
 Myelopathy/Tropical Spastic Paraparesis
 Patients by In Situ Hybridization
 Tanya J. Lebky, MD, Cecil H. Fox, PhD,
 Scott Koenig, MD, PhD, Michael C. Levin, MD,
 Nick Flerlage, BS, Shuji Izumo, MD, Elichi Sato, MD,
 Cedric S. Raine, PhD, Mitsubiro Osame, MD,
 and Steven Jacobson, PhD
- 176 Analysis of the SCA1 CAG Repeat in a Large Number of Families with Dominant Ataxia: Clinical and Molecular Correlations O. Dubourg, MD, A. Dürr, MD, G. Cancel, MS, G. Stevanin, MS, H. Chneiweiss, MD, PhD, C. Penet, BS, Y. Agid, MD, PhD, and A. Brice, MD
- 181 Changes in Excitability of Motor Cortical Circuitry in Patients with Parkinson's Disease M. C. Ridding, MSc, R. Inzelberg, MD, and J. C. Rothwell, PhD

- 189 Cerebrospinal Fluid 5-Hydroxyindoleacetic Acid and Homovanillic Acid in the Pediatric Opsoclonus-Myoclonus Syndrome M. R. Pranzatelli, MD, Y. Huang, MS, E. Tate, FNP, C, MN, M. Stanley, PhD, M. J. Noetzel, MD, S. M. Gospe, Jr, MD, PhD, and K. Banasiak, MD
- Multiple Sclerosis: Limited Diversity of the V82-J83 T-Cell Receptor in Chronic Active Lesions
 L. Battistini, MD, K. Selmaj, MD, PhD,
 C. Kowal, PhD, J. Ohmen, PhD, R. L. Modlin, MD,
 C. S. Raine, PhD, DSc, and C. F. Brosnan, PhD
- 204 Differentiation of Multiple System Atrophy from Idiopathic Parkinson's Disease Using Proton Magnetic Resonance Spectroscopy C. A. Davie, MRCP, G. K. Wenning, MD, G. J. Barker, PhD, P. S. Tofts, D Phil, B. E. Kendall, FRCR, N. Quinn, MD, W. I. McDonald, PhD, C. D. Marsden, DSc, and D. H. Miller, MD
- 211 Multiple, Diverse Senile Plaque-associated Proteins Are Ligands of an Apolipoprotein E Receptor, the α₂-Macroglobulin Receptor/ Low-Density-Lipoprotein Receptor-related Protein G. William Rebeck, PhD, Steven D. Harr, BA, Dudley K. Strickland, PhD, and Bradley T. Hyman, MD, PhD
- 218 Huntington's Disease Gene: Regional and Cellular Expression in Brain of Normal and Affected Individuals

 G. Bernbard Landwebrmeyer, MD,
 Sandra M. McNeil, PhD, Leon S. Dure IV, MD,
 Pei Ge, EdM, Hitoshi Aizawa, MD,
 Qin Huang, MD, PhD, Christine M. Ambrose, PhD,
 Mabel P. Duyao, PhD, Edward D. Bird, MD,
 Ernesto Bonilla, MD, PhD, Margot de Young, MD,
 Alejandro J. Avila-Gonzales, MD,
 Nancy S. Wexler, PhD, Marian DiFiglia, PhD,
 James F. Gusella, PhD, Marcy E. MacDonald, PhD,
 John B. Penney, MD, Anne B. Young, MD, PhD,
 and Jean-Paul Vonsattel, MD
- 231 Acute Human Stroke Studied by Whole Brain Echo Planar Diffusion-weighted Magnetic Resonance Imaging Steven Warach, MD, PhD, Jochen Gaa, MD, Bettina Siewert, MD, Piotr Wielopolski, PhD, and Robert R. Edelman, MD
- 242 Apolipoprotein E Genotypes in Parkinson's Disease with and without Dementia William C. Koller, MD, PhD, Sander L. Glatt, MD, Jean P. Hubble, MD, Anthony Paolo, PhD, Alexander I. Tröster, PhD, Michael S. Handler, MD, Rebecca T. Horvat, PhD, Coleman Martin, BS, Karen Schmidt, BS, Annette Karst, RN, Ellen M. Wijsman, PhD, Chang-En Yu, PhD, and Gerard D. Schellenberg, PhD

- 246 Topical Lidocaine Gel Relieves Postherpetic Neuralgia Michael C. Rowbotham, MD, Pamela S. Davies, RN, MS, and Howard L. Fields, MD, PhD
- 254 Apolipoprotein E and Alzheimer's Disease: Ethnic Variation in Genotypic Risks Gladys Maestre, MD, Ruth Ottman, PhD, Yaakov Stern, PhD, Barry Gurland, MD, Michael Chun, MD, Ming-Xin Tang, PhD, Michael Shelanski, MD, PhD, Benjamin Tycko, MD, PhD, and Richard Mayeux, MD, MSE
- 260 Serum Anti-GQ_{1b} IgG Antibodies Recognize Surface Epitopes on Campylobacter jejuni from Patients with Miller Fisher Syndrome Bart C. Jacobs, MD, Hubert Ph. Endtz, MD, Frans G. A. van der Meché, MD, Maarten P. Hazenberg, PhD, Herbert A. M. Achtereekte, MD, and Pieter A. van Doorn, MD
- 265 Plasticity of Nerve Afferents to Nigrostriatal Neurons in Parkinson's Disease Philippe Anglade, PhD, Shigeru Tsuji, PhD, France Javoy-Agid, PhD, Yves Agid, MD, PhD, and Etienne C. Hirsch, PhD

Brief Communications

- 273 Molecular and Clinical Findings in a Family with Dentatorubral-Pallidoluysian Atrophy Nicholas T. Potter, PhD, Michael A. Meyer, MD, Andrew W. Zimmerman, MD, Michael L. Eisenstadt, MD, PhD, and Ilse J. Anderson, MD
- 277 Concentrations of Amyloid β Protein in Cerebrospinal Fluid of Patients with Alzheimer's Disease W. A. van Gool, M. A. Kuiper, G. J. M. Walstra, E. Ch. Wolters, and P. A. Bolhuis
- 279 Proton Magnetic Resonance Spectroscopic Imaging in Patients with Frontal Lobe Epilepsy Paul A. Garcia, MD, Kenneth D. Laxer, MD, Jeroen van der Grond, PhD, James W. Hugg, PhD, Gerald B. Matson, PhD, and Michael W. Weiner, MD

Letters

- 282 No Protective Effect of Apolipoprotein E €2 Allele in Dutch Hereditary Cerebral Amyloid Angiopathy J. Haan, PhD, R. A. C. Roos, PhD, and E. Bakker, PhD
- 282 Supplementary Motor Area Activation Preceding Externally Triggered Movements J. A. Obeso, MD, J. L. Labres, MD, F. Barajas, MD, and E. Enriquez, MD
- 283 Reply Scott T. Grafton, MD
- 284 Reply
 P. Remy, MD, M. Zilbovicius, MD,
 and Y. Samson, MD

NO 3 MARCH 1995

Editorials

- 285 Episodic Ataxias as Channelopathies Robert C. Griggs, MD, and John G. Nutt, MD
- 287 Evidence that Aβ42 Is the Real Culprit in Alzheimer's Disease Steven G. Younkin, MD. PhD

Original Articles

- 289 A Gene for Hereditary Paroxysmal Cerebellar Ataxia Maps to Chromosome 19p Katayoun Vahedi, MD, Anne Joutel, MD, Patrick Van Bogaert, MD, Anne Ducros, MD, Jacqueline Maciaceck, Jean François Bach, MD, DSc, Marie Germaine Bousser, MD, and Elisabeth Tournier-Lasserve, MD
- 294 Amyloid β Protein (Aβ) Deposition: Aβ42(43) Precedes Aβ40 in Down Syndrome Takeshi Iwatsubo, MD, David M. A. Mann, PhD, Asano Odaka, MSc, Nobubiro Suzuki, PhD, and Yasuo Ibara, MD
- 300 Damage to Dopamine Systems Differs Between Parkinson's Disease and Alzheimer's Disease with Parkinsonism Angela M. Murray, PhD, Fredric B. Weihmueller, PhD, John F. Marshall, PhD, Howard I. Hurtig, MD, Gary L. Gottleib, MD, MA, and Jeffrey N. Joyce, PhD
- 313 Experimental Pneumococcal Meningitis:
 Cerebrovascular Alterations, Brain Edema, and
 Meningeal Inflammation Are Linked to the
 Production of Nitric Oxide
 Uwe Koedel, MD, Andrea Bernatowicz, PhD,
 Robert Paul, MD, Karl Frei, PhD,
 Adriano Fontana, MD, and Hans-Walter Pfister, MD
- 324 Leukoencephalopathy with Swelling and a Discrepantly Mild Clinical Course in Eight Children M. S. van der Knaap, MD, PhD, P. G. Barth, MD, PhD, H. Stroink, MD, O. van Nieuwenhuizen, MD, PhD, W. F. M. Arts, MD, PhD, F. Hoogenraad, MSc, and J. Valk, MD, PhD
- 335 Reduced Expression of Preproenkephalin in Striatal Neurons from Huntington's Disease Patients

 Eric K. Richfield, MD, PhD,

 Kathleen A. Maguire-Zeiss, PhD,

 Christopher Cox, PhD, Jennifer Gilmore, BS,
 and Pieter Voorn, PhD
- 344 Detection of Silent Cerebellar Lesions by Increasing the Inertial Load of the Moving Hand M. Manto, MD, E. Godaux, MD, and J. Jacquy, MD
- 351 Positron Emission Tomography and Magnetic Resonance Spectroscopy of Cerebral Glycosis in Children with Congenital Lactic Acidosis D. B. Duncan, MD, K. Herholz, MD, H. Kugel, PhD, B. Roth, MD, W. Ruitenbeek, PhD, W. Heindel, MD, K. Wienbard, PhD, and W.-D. Heiss, MD

- 359 Early-Onset Ataxia with Cardiomyopathy and Retained Tendon Reflexes Maps to the Friedreich's Ataxia Locus on Chromosome 9q Francisco Palau, MD, PhD, Giuseppe De Michele, MD, Juan J. Vilchez, MD, PhD, Massimo Pandolfo, MD, Eugènia Monrós, MSc, Sergio Cocozza, MD, Patricia Smeyers, MD, José Lopez-Arlandis, MD, PhD, Giuseppe Campanella, MD, Stefano Di Donato, MD, and Alessandro Filla. MD
- 363 Overactive Prefrontal and Underactive Motor Cortical Areas in Idiopathic Dystonia A. O. Ceballos-Baumann, MD, R. E. Passingham, PhD, T. Warner, MD, E. D. Playford, MD, C. D. Marsden, FRS, and D. J. Brooks, MD
- 373 Human Immunodeficiency Virus Type 1 Tat Activates Non-N-Methyl-D-Aspartate Excitatory Amino Acid Receptors and Causes Neurotoxicity

 David S. K. Magnuson, PhD, Bodo E. Knudsen, BSc, Jonathan D. Geiger, PhD,
 Robert M. Brownstone, MD, PhD,
 and Avindra Nath, MD
- 381 In Vitro Evidence for a Dual Role of Tumor Necrosis Factor-α in Human Immunodeficiency Virus Type 1 Encephalopathy Susan G. Wilt, PhD, Elizabeth Milward, PhD, Jia Min Zhou, MS, Kunihiko Nagasato, MD, Heather Patton, BS, Ray Rusten, Diane E. Griffin, MD, PhD, Michael O'Connor, MD, and Monique Dubois-Dalcq, MD
- 395 JC Virus DNA in Cerebrospinal Fluid of Human Immunodeficiency Virus-infected Patients: Predictive Value for Progressive Multifocal Leukoencephalopathy Dawn McGuire, MD, Steven Barbite, MS, Harry Hollander, MD, and Michael Miles, MD, PhD

Brief Communications

- 400 A New Mitochondrial DNA Mutation Associated with Progressive Dementia and Chorea: A Clinical, Pathological, and Molecular Genetic Study I. Nelson, PhD, M. G. Hanna, MRCP, N. Alsanjari, MRCPath, F. Scaravilli, FRCPath, J. A. Morgan-Hughes, FRCP, and A. E. Harding, FRCP
- 403 An Allelic Association Study of Monoamine Oxidase B in Parkinson's Disease S. L. Ho, MRCP, A. L. Kapadi, D. B. Ramsden, PhD, and A. C. Williams, MD, FRCP
- 405 The Gene for Hereditary Progressive Dystonia with Marked Diurnal Fluctuation Maps to Chromosome 14q H. Tanaka, MD, K. Endo, MD, S. Tsuji, MD, PhD, T. G. Nygaard, MD, D. E. Weeks, PhD, Y. Nomura, MD, and M. Segawa, MD

408 Hyperkalemic Periodic Paralysis with Cardiac Dysrhythmia: A Novel Sodium Channel Mutation?

Laime I. Baguero MD. Ricardo A Avala MD.

Jaime L. Baquero, MD, Ricardo A. Ayala, MD, Jianzhou Wang, MD, Richard G. Curless, MD, W. Gregory Feero, Eric P. Hoffman, PhD, and Makram R. Ebeid, MD

Letters

- 412 Dementia and Myoclonus: Differential Diagnosis of Early-Onset Alzheimer's Disease Samuel F. Berkovic, MD, Michel Melanson, MD, and Frederick Andermann, MD
- 412 Failure of Cytarabine/Interferon Therapy in Progressive Multifocal Leukoencephalopathy W. Heide, MD, D. Kömpf, MD, E. Reusche, MD, M. Bodemer, and T. Weber, MD

- 413 Corrective Saccades in Cerebellar Dysmetria
 A. M. Bronstein, MD, PhD, J. Shallo-Hoffman, PhD,
 R. Kanayama, MD, and P. Rudge, FRCP
 - 414 Reply
 Bertrand M. Gaymard, MD, Sophie Rivaud,
 Pierre Amarenco, MD,
 and Charles Pierrot-Deseilligny, MD
 - 414 Increased Levels of τ Protein in Cerebrospinal Fluid of Patients with Alzheimer's Disease—Correlation with Degree of Cognitive Impairment Christoph Hock, MD, Sidonie Golombowski, PhD, Werner Naser, PhD, and Franz Müller-Spahn, MD

Regular Departments

416 Books

NO 4 APRIL 1995

Editorials

- 419 The Multiple Sclerosis Lesion Henry F. McFarland, MD
- 421 Sodium Channel Blockade by Antibodies: A New Mechanism of Neurological Disease? Stephen G. Waxman, MD, PhD

Original Articles

- 424 The Adhesion Molecule and Cytokine Profile of Multiple Sclerosis Lesions

 Barbara Cannella, PhD,
 and Cedric S. Raine, PhD, DSc, FRCPath
- 436 Antibodies Against GM₁ Ganglioside Affect K⁺ and Na⁺ Currents in Isolated Rat Myelinated Nerve Fibers

 Tomoko Takigawa, MD, Hitoshi Yasuda, MD, PhD, Ryuichi Kikkawa, MD, PhD, Yukio Shigeta, MD, PhD, Takahiko Saida, MD, PhD, and Hiroshi Kitasato, MD, PhD
- 443 Deficits of Smooth-Pursuit Eye Movement After Unilateral Frontal Lobe Lesions Mark J. Morrow, MD, and James A. Sharpe, MD
- 452 A Clinical and Molecular Genetic Study of Dentatorubropallidoluysian Atrophy in Four European Families T. T. Warner, MRCP, L. D. Williams, BSc, R. W. H. Walker, FRCP, F. Flinter, MRCP, S. A. Robb, MRCP, S. E. Bundey, FRCP, M. Honavar, MRCPath, and A. E. Harding, FRCP
- 460 Pontine Ischemic Rarefaction
 Patrick Pullicino, MD, PhD, Peter Ostrow, MD, PhD,
 Lucia Miller, MD, Wendy Snyder, MD,
 and Frederick Munschauer, MD

- 467 Activated T Cells of Nonneural Specificity Open the Blood-Nerve Barrier to Circulating Antibody J. D. Pollard, PhD, FRACP, K. W. Westland, MSc, G. K. Harvey, PhD, S. Jung, MD, J. Bonner, J. M. Spies, MB, BS, K. V. Toyka, MD, and H.-P. Hartung, MD
- 476 Intrinsic Epileptogenicity of Human Dysplastic Cortex as Suggested by Corticography and Surgical Results

 André Palmini, MD, Antonio Gambardella, MD, Frederick Andermann, MD, FRCP(C), Francois Dubeau, MD, FRCP(C), Jaderson C. da Costa, MD, PhD, André Olivier, MD, PhD, FRCS(C), Donatella Tampieri, MD, Pierre Gloor, MD, PhD, Felipe Quesney, MD, PhD, Eva Andermann, MD, PhD, Eduardo Paglioli, MD, Eliseu Paglioli-Neto, MD, Ligia Coutinho, MD, PhD, Richard Leblanc, MD, FRCS(C), and Hyoung-Ihl Kim, MD, PhD
- 488 Cerebral Oxygen Supply and Utilization During
 Infant Cardiac Surgery
 Adre J. du Plessis, MD, Jane Newburger, MD,
 Richard A. Jonas, MD, Paul Hickey, MD,
 Hiroo Naruse, MD, Miles Tsuji, MD,
 Amy Walsh RN, BSN, Gene Walter, BS,
 David Wypij, PhD, and Joseph J. Volpe, MD
- 498 Basement Membrane Reduplication and Pericyte Degeneration Precede Development of Diabetic Polyneuropathy and Are Associated with Its Severity Caterina Giannini, MD, and Peter J. Dyck, MD

505 Effects of Brain-derived Neurotrophic Factor on Motor Dysfunction in Wobbler Mouse Motor Neuron Disease Ken Ikeda, MD, Bogdan Klinkosz, DVM, Tom Greene, PhD, Jesse M. Cedarbaum, MD.

Vivien Wong, PhD, Ronald M. Lindsay, PhD, and Hiroshi Mitsumoto, MD

- 512 Cerebrospinal Fluid Levels of Amyloid β-Protein in Alzheimer's Disease: Inverse Correlation with Severity of Dementia and Effect of Apolipoprotein E Genotype Roger M. Nitsch, MD, PhD, G. William Rebeck, PhD, Meihua Deng, MS, U. Ingrid Richardson, PhD, Marsha Tennis, RN, Dale B. Schenk, PhD, Carmen Vigo-Pelfrey, PhD, Ivan Lieberburg, MD, PhD, Richard J. Wurtman, MD, Bradley T. Hyman, MD, PhD, and John H. Growdon, MD
- 519 Active and Passively Induced Experimental Autoimmune Encephalomyelitis in Common Marmosets: A New Model for Multiple Sclerosis Luca Massacesi, MD, Claude P. Genain, MD, David Lee-Parritz, DVM, Norman L. Letvin, MD, Donald Canfield, DVM, and Stephen L. Hauser, MD
- The Role of Hypoventilation in a Sheep Model of Epileptic Sudden Death S. Claiborne Johnston, Jan K. Horn, John Valente, and Roger P. Simon

538 Selectively Distributed Processing of Visual Object Recognition in the Temporal and Frontal Lobes of the Human Brain M. Seeck, MD, D. Schomer, MD. N. Mainwaring, J. Ives, BSc, D. Dubuisson, MD, PhD, H. Blume, MD, PhD. R. Cosgrove, MD, B. J. Ransil, MD, and M.-M. Mesulam, MD

Brief Communications

- 546 Decreased CD3-mediated Interferon-y Production in Relapsing-Remitting Multiple Staley A. Brod, MD, Mohammed Khan, BS, John Bright, PhD, Subramaniam Sriram, MD, Gailen D. Marshall, Jr., MD, PhD, Evelyn M. Henninger, BA, Ronald H. Kerman, PhD, and Jerry S. Wolinsky, MD
- 550 Visual System Abnormalities in Adrenomyeloneuropathy Peter W. Kaplan, MB, FRCP, Bernd Kruse, MD. Ronald J. Tusa, MD, PhD, Jan Shankroff, MAS, JoEllen Rignani, BA, and Hugo W. Moser, MD

NO 5 MAY 1995

Editorials

- 553 Multiple Systems Atrophy and Nonfamilial Olivopontocerebellar Atrophy Are the Same Disease John B. Penney, Jr, MD
- 555 New Approaches to Locomotor Rehabilitation in Spinal Cord Injury Serge Rossignol, MD, PhD, and Hugues Barbeau, PT, PhD

Neurological Progress

557 Cortical Control of Saccades Charles Pierrot-Deseilligny, MD, Sophie Rivaud, Bertrand Gaymard, MD, René Müri, MD, and Anne-Isabelle Vermersch, MD

Original Articles

568 Positron Emission Tomography Studies on the Dopaminergic System and Striatal Opioid Binding in the Olivopontocerebellar Atrophy Variant of Multiple System Atrophy J. O. Rinne, MD, D. J. Burn, MD, MRCP, C. J. Mathias, DPhil, FRCP, N. P. Quinn, MD, FRCP, C. D. Marsden, MD, FRS, DSc, and D. J. Brooks, MD, FRCP

- 574 Locomotor Capacity of Spinal Cord in Paraplegic **Patients** V. Dietz, MD, G. Colombo, DM, L. Jensen, DM, and L. Baumgartner
- 583 Outcome after Spinal Reirradiation for Malignant Epidural Spinal Cord Compression David Schiff, MD, Edward G. Shaw, MD, and Terrence L. Cascino. MD
- Increased Risk of Mortality in Alzheimer's Disease Patients with More Advanced **Educational and Occupational Attainment** Yaakov Stern, PhD, Ming Xi Tang, PhD, Jean Denaro, MA, and Richard Mayeux, MD, MSE
- 596 Gene Dose of the ε4 Allele of Apolipoprotein E and Disease Progression in Sporadic Late-Onset Alzheimer's Disease Giovanni B. Frisoni, MD, Stefano Govoni, PhD. Cristina Geroldi, MD, Angelo Bianchetti, MD, Laura Calabresi, PhD, Guido Franceschini, PhD, and Marco Trabucchi, MD
- 605 The Apolipoprotein Ε ε2 Allele Is Associated with an Increased Risk of Early-Onset Alzheimer's Disease and a Reduced Survival Cornelia M. van Duijn, PhD, Peter de Knijff, PhD, Anita Wehnert, Joke De Voecht, Juliana B. Bronzova, MD, Louis M. Havekes, PhD, Albert Hofman, MD, PhD and Christine Van Broeckhoven, PhD

- 611 The Effect of Interferon-β on Blood-Brain Barrier Disruptions Demonstrated by Contrast-enhanced Magnetic Resonance Imaging in Relapsing-Remitting Multiple Sclerosis

 Lael A. Stone, MD, Joseph A. Frank, MD, Paul S. Albert, PhD, Craig Bash, MD, Mary E. Smith, MD, Heidi Maloni, RN, and Henry F. McFarland, MD
- 620 Ocular Neuromyotonia: Clinical Features, Physiological Mechanisms, and Response to Therapy Elliot M. Frohman, MD, PhD, and David S. Zee, MD
- 627 Acetylcholine Release in Myasthenia Gravis: Regulation at Single End-Plate Level Jaap J. Plomp, MSc, Gertrudis Th. H. Van Kempen, BSc, Marc H. De Baets, MD, Yvo M. F. Graus, PhD, Jan B. M. Kuks, MD, and Peter C. Molenaar, PhD
- 637 X-Linked Vacuolated Myopathy: Complement Membrane Attack Complex on Surface Membrane of Injured Muscle Fibers Marcello Villanova, MD, Jean Pierre Louboutin, MD, Danielle Chateau, MSc, Bruno Eymard, MD, Marguerite Sagniez, MD, Fernando M. S. Tomé, MD, and Micbel Fardeau, MD
- 646 Hereditary Ceruloplasmin Deficiency with Hemosiderosis: A Clinicopathological Study of a Japanese Family Hiroshi Morita, MD, Shu-ichi Ikeda, MD, Kanji Yamamoto, MD, Sayuri Morita, MD, Kunihiro Yoshida, MD, Shozo Nomoto, PhD, Masahiro Kato, MD, and Nobuo Yanagisawa, MD
- 657 Cognitive Scores in Carriers of Huntington's Disease Gene Compared to Noncarriers Tatiana Foroud, PhD, Eric Siemers, MD, Dawn Kleindorfer, BS, Danield J. Bill, EdD, M. E. Hodes, MD, PhD, James A. Norton, PhD, P. Michael Conneally, PhD, and Joe C. Christian, MD, PhD
- 665 Retinotopic and Directional Defects in Motion Discrimination in Humans with Cerebral Lesions Jason J. S. Barton, MD, FRCPC, James A. Sharpe, MD, FRCPC, and Jane E. Raymond, PhD

Brief Communications

- 676 Variance of Age at Onset in a Japanese Family with Amyotrophic Lateral Sclerosis Associated with a Novel Cu/Zn Superoxide Dismutase Mutation

 Masashi Aoki, MD, Koji Abe, MD, Kouji Houi, MD, Masahito Ogasawara, MD, Yoichi Matsubara, MD, Takaaki Kobayashi, PhD, Soichiro Mochio, MD, Kuniaki Narisawa, MD, and Yasuto Itoyama, MD
- Intracerebral Involvement in Scleroderma en Coup de Sabre: Report of a Case with Neuropathologic Findings

 Marilda H. Chung, MD, John Sum, MD,

 Martha J. Morrell, MD,

 and Dikran S. Horoupian, MD

Letters

- 682 Effect of Radiotherapy in Patients with Low-Grade Gliomas F. W. Kreth, MD, M. Peper, PhD, W. Bini, MD, and C. B. Ostertag, MD
- 682 Reply
 M. J. B. Taphoorn, MD
- 683 Treatment of Guillain-Barré Syndrome with Intravenous Methylprednisolone Richard Hughes, FRCP, and Anthony Swan, PhD
- 683 Reply
 L. H. Visser and F. G. A. van der Meché
- 684 Mechanism of Action of 4-Aminopyridine in the Symptomatic Treatment of Multiple Sclerosis Floyd A. Davis, MD, Dusan Stefoski, MD, and Fred N. Quandt, PhD
- 685 Increased Risk of Parkinson's Disease in Relatives of Patients Peter Vieregge, MD, and Ilse Heberlein, MD
- 685 Reply Haydeh Payami, MD, PhD, and John Nutt, MD
- 685 Parkinsonism Unmasked by Lovastatin Th. Müller, W. Kuhn, D. Pöhlau, and H. Przuntek

Regular Departments

687 Corrections

NO 6 JUNE 1995

Neurological Progress

691 Lyme Neuroborreliosis
Juan Carlos Garcia-Monco, MD, and
Jorge L. Benach, PhD

Original Articles

- 703 Magnetic Stimulation over the Cerebellum in Humans Yoshikazu Ugawa, MD, Yoshikazu Uesaka, MD, Yasuo Terao, MD, Ritsuko Hanajima, MD, and Ichiro Kanazawa, MD
- 714 Low Platelet Mitochondrial Complex I and Complex II/III Activity in Early Untreated Parkinson's Disease
 Richard H. Haas, MB, BChir,
 Fatemeh Nasirian, PhD, Kazutoshi Nakano, MD, David Ward, MB, ChB, Mary Pay, RN,
 Robert Hill, PhD, and Clifford W. Shults, MD
- 723 Recovery from Wernicke's Aphasia: A Positron Emission Tomographic Study
 Cornelius Weiller, MD, Christian Isensee, MD, Michel Rijntjes, MD, Walter Huber, MD, Stefan Müller, MD, Dirk Bier, Klaus Dutschka, Robert P. Woods, MD, Johannes Noth, MD, and Hans Christoph Diener, MD

- 733 First Seizures Associated with Playing Electronic Screen Games: A Community-based Study in Great Britain J. A. Quirk, FRACP, D. R. Fish, FRCP, S. J. M. Smith, MRCP, J. W. A. S. Sander, MD, S. D. Shorvon, FRCP, and P. J. Allen, MSc
- 738 Comparison of Ictal SPECT and Interictal PET in the Presurgical Evaluation of Temporal Lobe Epilepsy
 Susan S. Ho, FRACP, MRCP(UK),
 Samuel F. Berkovic, MD, FRACP,
 Salvatore U. Berlangieri, FRACP,
 Mark R. Newton, MD, FRACP, Gary F. Egan, PhD,
 Henri J. Tochon-Danguy, PhD, and
 W. John McKay, FRACP
- 746 Motor Reorganization in Acquired Hemidystonia
 A. O. Ceballos-Baumann, MD,
 R. E. Passingham, PhD, C. D. Marsden, FRS, and D. J. Brooks, MD
- 758 Expression of Heme Oxygenase-1 in the Senescent and Alzheimer-diseased Brain H. M. Schipper, MD, PhD, FRCP(C), S. Cissé, PhD, and E. G. Stopa, MD
- 769 Dentatorubral-Pallidoluysian Atrophy: Clinical Features Are Closely Related to Unstable Expansions of Trinucleotide (CAG) Repeat Takeshi Ikeuchi, MD, Reiji Koide, MD, Hajime Tanaka, MD, Osamu Onodera, MD, Shuichi Igarashi, MD, Hitoshi Takahashi, MD, Rui Kondo, MD, Atsushi Ishikawa, MD, Akemi Tomoda, MD, Teruhisa Miike, MD, Keiko Sato, MD, Yuetsu Ibara, MD, Toshiyuki Hayabara, Fumiko Isa, MD, Hitoshi Tanabe, MD, Susumu Tokiguchi, MD, Masataka Hayashi, MD, Natsue Shimizu, MD, Fusahiro Ikuta, MD, Haruhiko Naito, MD, and Shoji Tsuji, MD
- 776 Pallidotomy Increases Activity of Motor Association Cortex in Parkinson's Disease: A Positron Emission Tomographic Study Scott T. Grafton, MD, Cheryl Waters, MD, James Sutton, MD, Mark F. Lew, MD, and William Couldwell, MD

- 784 The Vasculopathy of Varicella-Zoster Virus Encephalitis Catherine Amlie-Lefond, MD, Bette K. Kleinschmidt-DeMasters, MD, Ravi Mahalingam, PhD, Larry E. Davis, MD, and Donald H. Gilden, MD
- 791 Movement-related Cortical Potentials Preceding Repetitive and Random-Choice Hand Movements in Parkinson's Disease T. Touge, MD, K. J. Werhahn, MD, J. C. Rothwell, PhD, and C. D. Marsden, MD
- 800 Interleukin-6 and Interleukin-1 Receptor Antagonist in Acute Stroke Nancy B. Beamer, MS, Bruce M. Coull, MD, Wayne M. Clark, MD, J. Samantha Hazel, BS, and Jenny R. Silberger, BA

Brief Communications

- 806 Common Variable Immunodeficiency and Inclusion Body Myositis: A Distinct Myopathy Mediated by Natural Killer Cells Marinos C. Dalakas, MD, and Isabel Illa, MD
- 810 Encephalopathy with Parkinsonian Features in Children Following Bone Marrow Transplantations and High-Dose Amphotericin B

 Stephen H. Mott, MD, Roger J. Packer, MD, L. Gilbert Vezina, MD, Sudesh Kapur, MD, Patricia A. Dinndorf, MD, Joan A. Conry, MD, Michael R. Pranzatelli, MD, and Ralph R. Quinones, MD
- 815 Cephalosporin-induced Recurrent Aseptic Meningitis George B. Creel, MD, and Mark Hurtt, MD
- 817 Studies of the Coding Region of the Neuronal Glutamate Transporter Gene in Amyotrophic Lateral Sclerosis T. Meyer, U. Lenk, G. Küther, A. Weindl, A. Speer, and A. C. Ludolph

Indexes to Volume 37

- 822 Author Index
- 825 Subject Index

Regular Departments

820 Books

SUPPLEMENT 1 TO VOLUME 37

Clinical Neuroimmunology and Immunotherapy

- S1 Introduction Marinos C. Dalakas, MD S2 Basic Aspects of Neuroimmunology as They Relate to Immunotherapeutic Targets: Present and Future Prospects Marinos C. Dalakas, MD S14 Guillain-Barré Syndrome and Chronic Inflammatory Demyelinating Polyneuropathy: Immune Mechanisms and Update on **Current Therapies** F. G. A. van der Meché, MD, and P. A. van Doorn, MD S32 Pathogenesis and Therapy of Neuropathies Associated with Monoclonal Gammopathies Norman Latov, MD, PhD Chronic Motor Neuropathies: Diagnosis, Therapy, and Pathogenesis Andrew J. Kornberg, MB BS, FRACP, and Alan Pestronk, MD S51 Myasthenia Gravis: Immunological Mechanisms and Immunotherapy Richard A. Lewis, MD, James F. Selwa, MD, and Robert P. Lisak, MD
- S63 Lambert-Eaton Myasthenic Syndrome: Clinical Diagnosis, Immune-mediated Mechanisms, and Update on Therapies

 Donald B. Sanders, MD
- S74 Immunopathogenesis of Inflammatory Myopathies Marinos C. Dalakas, MD
- S87 Multiple Sclerosis: Immune Mechanism and Update on Current Therapies
 Shalini Bansil, MD, Stuart D. Cook, MD, and Christine Rohowsky-Kochan, PhD
- S102 Autoimmune Central Nervous System Paraneoplastic Disorders: Mechanisms, Diagnosis, and Therapeutic Options Edward J. Dropcho, MD
- S114 Cerebrovascular Disease with Antiphospholipid Antibodies: Immune Mechanisms, Significance, and Therapeutic Options Edward Feldmann, MD, and Steven R. Levine, MD
- S131 Neurological Manifestation of Vasculitis: Update on Immunopathogenic Mechanisms and Clinical Features Patricia M. Moore, MD

